In children with type 3 osteogenesis imperfecta and thoracic insufficiency syndrome, thoracic elongation surgery using a novel expandable spino-thoracic fixation device significantly improves pulmonary function, weight gain, and spinal deformities, without significant complications, according to a study published online Nov. 7 in Spine.

Leon Kaplan, M.D., of the Hadassah Hebrew University Medical Center in Jerusalem, and colleagues conducted a prospective series study to
examine outcomes of four children aged 8 to 12 years with type 3 OI and TIS who underwent surgery with a novel expandable spino-thoracic fixation device.

After an average follow-up of 24 months, the researchers found that the mean Cobb angle in the coronal plane was improved up to 32 percent. Pulmonary function improved for all patients, with clinically significant increases in forced vital capacity, forced expiratory flow, and partial pressure of oxygen values, and normalization of partial pressure of carbon dioxide values. Patient weight increased, with patients moving from below the third percentile to the third to seventh percentile at the six-month follow-up. All patients and their caregivers reported improved self-image and functional level.

"Thoracic elongation improved pulmonary function, facilitated weight gain, and made an indirect correction of spinal deformities (Cobb angle) by over 30 percent, with no significant complications," the authors write. "Due to the rarity of this condition, a multicenter study to gain experience in a larger number of patients is recommended."

One or more of the authors disclosed receipt of benefits from a commercial entity related directly or indirectly to this study.

More information: Abstract
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Citation: Novel device benefits type 3 osteogenesis imperfecta (2012, November 20) retrieved 7 October 2023 from https://medicalxpress.com/news/2012-11-device-benefits-osteogenesis-imperfecta.html